General

Guideline Title

EFNS guidelines on diagnosis and treatment of primary dystonias.

Bibliographic Source(s)

Albanese A, Asmus F, Bhatia KP, Elia AE, Elibol B, Filippini G, Gasser T, Krauss JK, Nardocci N, Newton A, Valls-Sole J. EFNS guidelines on diagnosis and treatment of primary dystonias. Eur J Neurol. 2011 Jan;18(1):5-18. PubMed

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Albanese A, Barnes MP, Bhatia KP, Fernandez-Alvarez E, Filippini G, Gasser T, Krauss JK, Newton A, Rektor I, Savoiardo M, Valls-Sole J. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. Eur J Neurol 2006 May;13(5):433-44.

Recommendations

Major Recommendations

The levels of evidence (class I-IV) supporting the recommendations and ratings of recommendations (A-C, Good Practice Point [GPP]) are defined at the end of the "Major Recommendations" field.

Diagnosis and Classification

1. The diagnosis of dystonia is clinical, the core being abnormal postures (with or without tremor) and the recognition of specific features, e.g., *gestes antagonistes*, overflow and mirror movements (GPP).
2. The classification of dystonia is important for providing appropriate management, prognostic information, genetic counselling and treatment (GPP).
3. Because of the lack of specific diagnostic tests, expert observation is recommended. Using a structured flow chart (Albanese & Lalli, 2009) may increase diagnostic accuracy (GPP).
4. Appropriate investigations are required if the initial presentation or the course suggests heredodegenerative or secondary (symptomatic) dystonia (GPP).
5. Assessment of dystonia should be performed using a validated rating scale (GPP).

Use of Genetic Test in Diagnosis and Counseling

1. Genetic testing should be performed after establishing the clinical diagnosis. Genetic testing is not sufficient to make a diagnosis of dystonia

without clinical features of dystonia (Bressman et al., 2000; "Points to consider," 1995; Klein et al., 1999) (Level B). Genetic counselling is recommended.
2. DYT1 testing is recommended for patients with limb-onset, primary dystonia with onset before age 30 (Klein et al., 1999) (Level B), as well as in those with onset after age 30 if they have an affected relative with early-onset dystonia (Bressman et al., 2000; Klein et al., 1999) (level B).
3. In dystonia families, DYT1 testing is not recommended in asymptomatic individuals (GPP).
4. DYT6 testing is recommended in early-onset dystonia or familial dystonia with cranio-cervical predominance (Djarmati et al., 2009; Bressman et al., 2009) or after exclusion of DYT1 (GPP).
5. A diagnostic levodopa trial is warranted in every patient with early-onset dystonia without an alternative diagnosis (Robinson et al., 1999) (GPP).
6. Individuals with early-onset myoclonus affecting the arms or neck, particularly if positive for autosomal-dominant inheritance and if triggered by action, should be tested for the DYT11 gene (Valente et al., 2005) (GPP). If direct sequencing of the SGCE gene is negative, gene dosage studies increase the proportion of mutation-positives (Level C).
7. Diagnostic testing for the paroxysmal non-kinesigenic form of dystonia (PNKD) gene (DYT8) is recommended in symptomatic individuals with PNKD (GPP).
8. Gene testing for mutation in GLUT1 is recommended in patients with paroxysmal exercise-induced dyskinesias, especially if involvement of GLUT1 is suggested by low cerebrospinal fluid (CSF)/serum glucose ratio, epileptic seizures or haemolytic anaemia (GPP).

Use of Neurophysiology in the Diagnosis and Classification of Dystonia

1. Neurophysiological tests are not routinely recommended for the diagnosis or classification of dystonia; however, multiple simultaneous electromyography (EMG) recordings from various muscles may contribute to the clinical assessment by showing characteristic features of dystonia (Albanese & Lalli, 2009) (GPP).

Use of Brain Imaging in the Diagnosis of Dystonia

1. Structural brain imaging is not routinely required when there is a confident diagnosis of primary dystonia in adult patients, because a normal study is expected in primary dystonia (Rutledge et al., 1988) (GPP).
2. Structural brain imaging (magnetic resonance imaging [MRI]) is necessary for screening of secondary forms of dystonia (Meunier et al., 2003) (GPP). Computed tomography may be required to differentiate between calcium and iron accumulation.
3. Pre-synaptic dopaminergic scan (dopamine transporter [DAT] or ^{18}F-DOPA) is useful to differentiate between dopa-responsive dystonia (DRD) and juvenile Parkinson's disease presenting with dystonia (GPP). This can also be useful to distinguish dystonic tremor from parkinsonian tremor (GPP).

Treatment

Botulinum Toxins (BoNT)

1. BoNT/A (or type B if there is resistance to type A) can be regarded as first line treatment for primary cranial (excluding oromandibular) or cervical dystonia (Costa et al., 2005; American Academy of Ophthalmology, 1989) (Level A).
2. BoNT/A is effective for writer's cramp (Kruisdijk et al., 2007) (Level A) and is possibly effective in other types of upper limb dystonia, but controlled dose adjustments are needed because of frequent muscle weakness (GPP).
3. BoNT/A is probably effective for adductor-type laryngeal dystonia, but there is insufficient evidence to support efficacy in abductor-type laryngeal dystonia and in muscular tension dysphonia (GPP).
4. BoNT are safe and efficacious when repeated treatments are performed over many years (GPP), but doctors and patients should be aware that excessive cumulative doses may be dangerous, particularly in children (GPP).
5. BoNT injections can be performed by direct inspection; EMG- or ultrasound-assisted targeting may improve clinical outcome (GPP).
6. BoNT should not be used in patients affected by a disorder of neuromuscular transmission or in presence of local infection at the injection site. The recommended dosage should not be exceeded (GPP).

Other Medical Treatments

No new class A or B data are available for oral medications. Therefore, the previously reported recommendations and good practice points are retained (Albanese et al., 2006).

Neurosurgical Procedures

Deep Brain Stimulation (DBS)

1. Pallidal DBS is considered a good option, particularly for primary generalized or segmental dystonia, after medication or BoNT has failed to provide adequate improvement (Kupsch et al., 2006) (Level A).
2. Pallidal DBS can be considered a good option for cervical dystonia, after medication or BoNT has failed to provide adequate improvement (Kiss et al., 2007) (Level B).
3. Pallidal DBS, in general, is less effective in secondary dystonia with the exception of tardive dystonia (Vidailhet et al., 2009; Gruber et al., 2009) (Level C).
4. This procedure requires a specialized expertise and a multidisciplinary team and is not without side effects (GPP).

Other Surgical Procedures

In the past 5 years, there have been no new studies providing class I or II evidence for selective peripheral denervation, myectomy and myotomy, intrathecal baclofen or radiofrequency lesioning. Therefore, the previously reported recommendations and Good Practice Points are retained (Albanese et al., 2006).

Physical Therapy and Rehabilitation

1. Transcutaneous electrical nerve stimulation to forearm flexor muscles administered is probably effective in patients with writer's cramp (Tinazzi et al., 2005) (Level B).
2. The task force encourages the conduction of new randomized controlled studies on these potentially useful interventions, particularly for patients with upper limb dystonia (GPP).

Definitions:

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
c. Exclusion/inclusion criteria are clearly defined
d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Point Where only class IV evidence was available but consensus could be achieved the task force has proposed Good Practice Points.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Primary (idiopathic) dystonia, including:

- Primary pure dystonia
- Primary plus dystonia
- Primary paroxysmal dystonia

Guideline Category

Diagnosis

Evaluation

Management

Treatment

Clinical Specialty

Family Practice

Internal Medicine

Medical Genetics

Neurology

Pediatrics

Intended Users

Physicians

Guideline Objective(s)

To provide a revised version of earlier guidelines published in 2006 on diagnosis and treatment of primary dystonias

Target Population

Patients with primary (idiopathic) dystonia

Interventions and Practices Considered

Diagnosis

1. Expert observation and neurological examination
2. Classification of dystonia (by cause, age at onset, and distribution)
3. Use of a validated rating scale for assessment of dystonia
4. Genetic testing and genetic counselling
5. Neurophysiological tests in selected cases
6. Diagnostic levodopa trial
7. Magnetic resonance imaging (MRI) for screening of secondary dystonia (or computed tomography when brain calcifications are suspected)
8. Pre-synaptic dopaminergic scan (dopamine transporter [DAT] or ^{18}F-DOPA)

Treatment

1. Botulinum toxin (BoNT)
2. Levodopa
3. Other oral medications
4. Neurosurgical procedures
    - Pallidal deep brain stimulation (DBS)
    - Selective peripheral denervation and myectomy
    - Intrathecal baclofen
    - Other surgical procedures
5. Transcutaneous electrical nerve stimulation to forearm flexor muscles

Major Outcomes Considered

- Utility and diagnostic accuracy of genetic testing
- Effectiveness of treatment in terms of severity and disability improvement and pain relief

Methodology

Methods Used to Collect/Select the Evidence

Hand-searches of Published Literature (Primary Sources)

Hand-searches of Published Literature (Secondary Sources)

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Computerized MEDLINE and EMBASE searches (2005-July 2009) were conducted using a combination of text words and MeSH and EMTREE terms 'dystonia', 'blepharospasm', 'torticollis', 'writer's cramp', 'Meige syndrome', 'dysphonia' and 'sensitivity and specificity' or 'diagnosis', and 'clinical trial' or 'random allocation' or 'therapeutic use' limited to human studies. The Cochrane Library and the reference lists of all known primary and review articles were searched for relevant citations. No language restrictions were applied.

Number of Source Documents

In addition to the previously published literature, 299 papers were found.

- Primary diagnostic studies - 191
- Efficacy studies - 108

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Evidence Classification Scheme for a Diagnostic Measure

Class I: A prospective study in a broad spectrum of persons with the suspected condition, using a 'gold standard' for case definition, where the test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class II: A prospective study of a narrow spectrum of persons with the suspected condition, or a well-designed retrospective study of a broad spectrum of persons with an established condition (by 'gold standard') compared to a broad spectrum of controls, where test is applied in a blinded evaluation, and enabling the assessment of appropriate tests of diagnostic accuracy

Class III: Evidence provided by a retrospective study where either persons with the established condition or controls are of a narrow spectrum, and where test is applied in a blinded evaluation

Class IV: Any design where test is not applied in blinded evaluation OR evidence provided by expert opinion alone or in descriptive case series (without controls)

Evidence Classification Scheme for a Therapeutic Intervention

Class I: An adequately powered prospective, randomized, controlled clinical trial with masked outcome assessment in a representative population or an adequately powered systematic review of prospective randomized controlled clinical trials with masked outcome assessment in representative populations. The following are required:

a. Randomization concealment
b. Primary outcome(s) is/are clearly defined
c. Exclusion/inclusion criteria are clearly defined
d. Adequate accounting for dropouts and crossovers with numbers sufficiently low to have minimal potential for bias
e. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences

Class II: Prospective matched-group cohort study in a representative population with masked outcome assessment that meets a–e above or a randomized, controlled trial in a representative population that lacks one criteria a–e

Class III: All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome assessment is independent of patient treatment

Class IV: Evidence from uncontrolled studies, case series, case reports, or expert opinion

Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review

Description of the Methods Used to Analyze the Evidence

Studies of diagnosis, diagnostic test, and various treatments for patients suffering from dystonia were considered and rated as level A to C according to the recommendations for European Federation of Neurological Societies (EFNS) scientific task forces (see the "Rating Scheme for the Strength of the Evidence" field).

Methods Used to Formulate the Recommendations

Expert Consensus (Consensus Development Conference)

Description of Methods Used to Formulate the Recommendations

The results of the literature searches were circulated by email to the task force members for comments. The task force chairman prepared a first draft of the manuscript based on the results of the literature review, data synthesis and comments from the task force members. The draft and the recommendations were discussed during a conference held in Florence on 12 September 2009, until consensus was reached within the task force.

Where only Class IV evidence was available but consensus could be achieved, the task force has proposed Good Practice Points.

Rating Scheme for the Strength of the Recommendations

Rating of Recommendations for a Diagnostic Measure

Level A rating (established as useful/predictive or not useful/predictive) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (established as probably useful/predictive or not useful/predictive) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (established as possibly useful/predictive or not useful/predictive) requires at least two convincing class III studies.

Rating of Recommendations for a Therapeutic Intervention

Level A rating (established as effective, ineffective, or harmful) requires at least one convincing class I study or at least two consistent, convincing class II studies.

Level B rating (probably effective, ineffective, or harmful) requires at least one convincing class II study or overwhelming class III evidence.

Level C rating (possibly effective, ineffective, or harmful) requires at least two convincing class III studies.

Good Practice Point Where only class IV evidence was available but consensus could be achieved the task force has proposed Good Practice Points.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Peer Review

Description of Method of Guideline Validation

The guidelines were validated according to the European Federation of Neurological Societies (EFNS) criteria (see the "Availability of Companion Documents" field in this summary).

Evidence Supporting the Recommendations

References Supporting the Recommendations

Albanese A, Barnes MP, Bhatia KP, Fernandez-Alvarez E, Filippini G, Gasser T, Krauss JK, Newton A, Rektor I, Savoiardo M, Valls-Sole J. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. Eur J Neurol. 2006 May;13(5):433-44. [111 references] PubMed

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Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for selected recommendations (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

Appropriate diagnosis and treatment of primary (idiopathic) dystonia

Potential Harms

- *Botulinum toxin (BoNT)*. The main side effects consisted in muscle weakness in or around the injected region. Immunogenicity was found to be low for BoTN/A in long-term use, although might be higher for BoTN/B. A meta-analysis performed on children with cerebral palsy

found that adverse events are more frequent amongst children with cerebral palsy than in individuals with other conditions. Occasional occurrences of botulism-like symptoms have been reported in children and in adults treated with BoNT products; therefore, the United States Food and Drug Administration has ordered the manufacturers to add a boxed warning to the prescribing information for each product about the potential for serious side effects at sites distant from injection. Furthermore, the possible occurrence of central effect following BoNT because of axonal migration and neuronal transcytosis has been recently suggested, but not unequivocally demonstrated. Two class I studies found that dry mouth and dysphagia were more frequent with BoNT/B treatment than with BoNT/A. A class II study reported that patients treated with BoNT/B had less saliva production and more severe constipation than those treated with BoNT/A.

- *Pallidal deep brain stimulation (DBS)*. In a sham-stimulation group, a total of 22 adverse events occurred in 19 patients (the most frequent adverse event was dysarthria) during an overall follow-up of 6 months. Safety aspects which have to be considered include surgery-related complications, stimulation-induced side effects and hardware-related problems. Recently, it was noted that *globus pallidus internus* (Gpi) DBS in patients with segmental dystonia may induce a parkinsonian gait or bradykinesia in extremities which were not affected by dystonia at chronic stimulation with high voltage. Chronic stimulation uses both higher pulse width and voltage than in Parkinson disease (PD), which results in much higher energy consumption and earlier battery depletion; replacement may be needed sometimes every 2 years or less. Sudden battery depletion may induce acute recurrence of dystonia, sometimes resulting in a medical emergency.

Contraindications

Contraindications

Botulinum toxin (BoNT) should not be used in patients affected by a disorder of neuromuscular transmission or in presence of local infection at the injection site.

Qualifying Statements

Qualifying Statements

This guideline provides the view of an expert task force appointed by the Scientific Committee of the European Federation of Neurological Societies (EFNS). It represents a peer-reviewed statement of minimum desirable standards for the guidance of practice based on the best available evidence. It is not intended to have legally binding implications in individual cases.

Implementation of the Guideline

Description of Implementation Strategy

The European Federation of Neurological Societies has a mailing list and all guideline papers go to national societies, national ministries of health, World Health Organisation, European Union, and a number of other destinations. Corporate support is recruited to buy large numbers of reprints of the guideline papers and permission is given to sponsoring companies to distribute the guideline papers from their commercial channels, provided there is no advertising attached.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

Bibliographic Source(s)

Albanese A, Asmus F, Bhatia KP, Elia AE, Elibol B, Filippini G, Gasser T, Krauss JK, Nardocci N, Newton A, Valls-Sole J. EFNS guidelines on diagnosis and treatment of primary dystonias. Eur J Neurol. 2011 Jan;18(1):5-18. PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2006 May (revised 2011 Jan)

Guideline Developer(s)

European Academy of Neurology - Medical Specialty Society

Source(s) of Funding

European Federation of Neurological Societies

Guideline Committee

European Federation of Neurological Societies Task Force on Diagnosis and Treatment of Primary Dystonias

Composition of Group That Authored the Guideline

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Financial Disclosures/Conflicts of Interest

Not stated

Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Albanese A, Barnes MP, Bhatia KP, Fernandez-Alvarez E, Filippini G, Gasser T, Krauss JK, Newton A, Rektor I, Savoiardo M, Valls-Sole J. A systematic review on the diagnosis and treatment of primary (idiopathic) dystonia and dystonia plus syndromes: report of an EFNS/MDS-ES Task Force. Eur J Neurol 2006 May;13(5):433-44.

Guideline Availability

Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies (EFNS) Web site .

Availability of Companion Documents

The following is available:

- Brainin M, Barnes M, Baron JC, Gilhus NE, Hughes R, Selmaj K, Waldemar G; Guideline Standards Subcommittee of the EFNS Scientific Committee. Guidance for the preparation of neurological management guidelines by EFNS scientific task forces – revised recommendations 2004. Eur J Neurol. 2004 Sep;11(9):577-81. Electronic copies: Available in Portable Document Format (PDF) from the European Federation of Neurological Societies Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI on March 26, 2007. The information was verified by the guideline developer on May 3, 2007. This summary was updated by ECRI Institute on May 26, 2009, following the U.S. Food and Drug Administration advisory on Botox, Botox Cosmetic (Botulinum toxin Type A), and Myobloc (Botulinum toxin Type B). This summary was updated by ECRI Institute on August 17, 2009, following the updated FDA advisory on Botox and Botox Cosmetic (Botulinum toxin Type A), and Myobloc (Botulinum toxin Type B). This NGC summary was updated by ECRI Institute on February 20, 2012.

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